ON

TUMORS OF THE OPTIC NERVE.

BY

H. KNAPP, M.D.,

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ON TUMORS OF THE OPTIC NERVE.

A VERY limited number of tumors of the optic nerve has hitherto been recorded. W. Goldzieher, in a very valuable paper on the subject, published in Graefe's Archiv für Ophthalmologie, 1873, states that in modern medical literature he has found only six cases of the kind, to which he himself adds three others. I have found records of a few additional cases, and have seen three myself, two having been under my own care and one under that of Dr. E. Gruening at the New York Ophthalmic and Aural Institute. I shall give a brief sketch of some of these cases, in order to obtain the data necessary for a general description of the affection.

CASE I.—(Described by A. von Graefe, in his Archives, vol. x. part 1, p. 193 etc., 1864.) A farmer, æt. 23, had first noticed, two years previously, diplopia and protrusion of his right eye. The exophthalmos increased steadily, but the diplopia subsequently disappeared from the sight of that eye becoming lost. No pain was felt until a few weeks before the patient was first seen, when secondary infiltration of the cornea set in. On examination, there was found exophthalmos of 9‴, in the direction of the orbital axis; the lids could not be closed; the mobility of the globe was restricted inward, but in other directions was free; the centre of motion coincided with that of the globe. Behind the globe, a soft, elastic tumor was felt; S. = 0. The retinal veins were tortuous and dilated, the arteries attenuated. The inner half of the optic disk was swollen, with abrupt, slightly overlapping borders, reddish, and concealing the bloodvessels; the outer half was level.

Graefe diagnosticated a relatively benign, orbital tumor, probably of a fibro-sarcomatous nature. From the fact that the centre of rotation corresponded with the centre of the globe, he concluded that a stratum of loose connective tissue separated the tumor from the globe. The freedom of the outer half of the optic disk led him to the supposition that the blindness was not caused by compression, but by the immediate transition of the tumor to the nerve, or possibly by its origin in the nerve itself. The tumor being situated within the muscular funnel, and surrounding the optic nerve, its extirpation with preservation of the eyeball, he said, could not be thought of. He enucleated the eyeball, slit the stratum of connective tissue in front of the morbid growth, laid the bluish-red looking tumor bare, and removed it without difficulty. The operation was followed by alarming cerebral symptoms, intense headache and nausea; the pulse fell from 80 to 52 beats, and the temperature rose to 40° C. (104° F.). These symptoms subsided upon the appearance of an extensive suppuration in the wound. The patient recovered.

The tumor, examined by Recklinghausen, was pear-shaped, a little larger than a pigeon's egg, partially soft and gelatinous, and partially fibrous. The bulk of the optic nerve lay as a compact cord on one side of the tumor, and was covered, together with the whole tumor, by a tough, fibrous capsule. The limit of the nerve which looked toward the tumor, was ill-defined. The fibres of the cord-like portion of the optic nerve were preserved in their whole course, the others entered the tumor and were lost in it. This condition was found

in both the cut ends of the nerve, that is at its entrance into and its emergence from the tumor. The growth therefore was a *myxoma* of the optic nerve, originating in the inner sheath and perineurium internum. The swelling of the inner half of the intra-ocular end of the optic nerve was caused (according to the examination of Schweigger) "by a proliferation of indifferent (lymphoid) cells, as might be expected in so young a formation."

Case II.—(Described by A. Rothmund, Zehender's Klin. Mon., 1863, S. 261.) The patient was a healthy looking girl of fifteen years. At the beginning of her second year, her left eye began to be pushed forward and downward, and was excessively painful. She could see with that eye. In the course of four or five months the eye became blind, but kept free from pain; yet the lids were swollen every now and then. From that time to her sixteenth year, she felt no annoyance from the eye, though it slowly but constantly grew larger. When seen by Dr. R., the orbit was occupied by a roundish, faintly fluctuating tumor, the size of a large hen's egg. The tumor was totally covered by congested conjunctiva, but incompletely by the lids, and moved harmoniously with the healthy eye in every direction. Remnants of the cornea were recognizable at its apex, which was very tender to the touch. The palpebral fissure was two and a half inches in length, and the orbit was greatly distended.

Dr. Rothmund removed the tumor in the manner of an ordinary enucleation. Its pedicle was severed near the optic foramen. In some weeks the orbital wound was closed by granulations. The patient recovered.

At the posterior end of the tumor a piece of the optic nerve, 3‴ in length, was preserved. The growth was a total degeneration of the optic nerve, consisting of fibrous tissue which inclosed smaller and larger cysts, the cavities of which were pervaded by a network of white, delicate fibres containing a vascular, gelatinous substance. The microscopic examination, made by Prof. Buhl, showed the characteristics of a *myxoma*, which is frequently met with in other nerves. The eyeball had been flattened by the tumor in such a way that the optic disk touched the cornea.

Case III.—(A. von Graefe, Arch. f. Ophth., x. 1, S. 201, 1864.) The patient was a young lady, æt. 24, who for two years had noticed a slight exophthalmos with amblyopia and contraction of the visual field of her left eye. With the ophthalmoscope Graefe discovered choked disk and spontaneous arterial pulsation. During the next five years the exophthalmos slowly increased, while sight was reduced to quantitative perception of light. The exophthalmos measured 8‴, and showed a marked deviation outward. Mobility preserved, but reduced in every direction. The centre of rotation corresponded approximately to the centre of the globe. A soft tumor was located immediately behind the eye, more on the nasal than on the temporal side. Between the upper and inner recti muscles it could be seen through the conjunctiva as a smooth, reddish-yellow intumescence. In the outer part of the field of vision, there was no perception of light. The optic disk was atrophic.

The patient was operated on by Langenbeck, who first enucleated the eyeball, and then the tumor, which did not extend to the apex of the orbit. It was as large as a pigeon's egg. The optic nerve passed as a flattened cord along its nasal side, but a considerable part of it entered the tumor and was lost in it. The posterior part of the eyeball was flattened by the pressure from the tumor.

The tumor, examined by Recklinghausen, was found to be a *myxo-sarcoma* of the optic nerve. Its anterior part contained a cyst, around and behind which there was a soft, partly gelatinous substance, consisting of very delicate, interlacing fibres which inclosed round and oval cells. The gelatinous liquid which could be pressed out of the tumor grew very opaque on addition of acetic acid. Larger portions of the tumor showed a substance almost entirely composed of cells, between which some larger bloodvessels ramified.

Case IV.—(Described by E. Neumann, in Archiv für Heilkunde, xiii. S.

310.) The patient, æt. 20, had suffered from headache for six years, and had noticed a protrusion of her eye for three years. The mobility of the eye was preserved, and was only a little restricted in an upward direction. The vision was almost normal. On palpation a hard immovable tumor was felt surrounding the posterior part of the globe on all sides. Dr. Jacobson, of Königsberg, removed the eye together with the tumor.

The tumor was three-quarters of an inch in length, the size of a walnut, and immediately behind the eye surrounded the optic nerve, which passed quite loosely across it. The neoplasm was connected with the outer sheath, which on section showed itself as a distinct, white border-line. The inner sheath was smooth and shining. Between the two sheaths, delicate cords were stretched when the nerve was drawn away from the tumor. The microscopic examination discovered that the structure of the tumor was partly compact, partly alveolar. The alveolar part had a stroma which resembled that of a cancer, inclosing accumulations of fusiform cells in concentric layers. The centres of many cells were incrusted. Teased preparations showed that these fusiform elements were flat endothelioid cells. The compact part of the tumor consisted of coarse fibrils, here and there inclosing numerous accumulations of sarcomatous cells. The periphery of the tumor consisted of adipose tissue. The tumor is styled by Neumann a *psammoma*, on account of the prevalence of the incrusted, arenoid bodies in the alveolar part of the tumor.

Case V.—(Described by H. Knapp, in Archives of Ophthalmology and Otology, vol. iv. pp. 323–354, 1875, with four plates.) Mrs. J. K., æt. 40, was first examined in August, 1871. Her left eye was normal in function and structure; the right protruded about 5‴ in a forward and slightly downward direction. The protrusion had begun six months previously, and the sight of that eye had gradually become impaired. She suffered from occasional headaches, which always increased the protrusion of the eye. The movements of the globe were somewhat restricted, chiefly in an upward direction. There was no pulsation, nor bruit. S. with +10 Sn. $\frac{20}{100}$; F. complete. The optic disk showed a steep elevation, arteries small, veins dilated and tortuous. The ophthalmoscopic condition did not materially change for three years. In June, 1874, a hardish tumor was felt, on palpation, on the inner and upper side of the posterior part of the globe. It moved with the eye. A free space was felt between the tumor and the orbital wall. The globe was dislocated slightly outward, and considerably downward (4‴–5‴), but mostly forward (6‴). Inversion was perfect, whereas the strongest eversion brought the corneal margin no nearer than 2‴ from the outer commissure; downward rotation was free, but upward rotation moved the eye no higher than the horizontal median plane; S. $\frac{10}{200}$. F. complete. Media clear, pupil responsive. Papilla raised, with abrupt borders, like a "jockey-cap." Venous hyperæmia marked. General health good. For the last years, great pain in eye and head. The diagnosis of orbital tumor could now be more specifically revised into that of a tumor of the optic nerve, and acting upon this supposition I attempted, on June 10, 1874, to enucleate the tumor while preserving the eyeball. I succeeded in the following manner:—

I opened the conjunctiva by a circular section between the inner and superior recti muscles, and laid the anterior portion of the tumor bare. Then I introduced my left forefinger into the wound, and under its guidance freed the hardish tumor from the soft, surrounding orbital tissue. After that, I dissected the mass from the posterior part of the globe, with which it was connected by a thin layer of connective tissue. Further, I severed the optic nerve, and then introduced my finger as deeply into the orbit as possible. Not being able to reach the apex of the tumor, which was close to the optic foramen, I cut the tumor across as near the summit of the orbit as I could reach. Now I rotated the growth on its antero-posterior axis, and severed such connections with its surroundings as had before escaped. All this was done with a pair of strabismus scissors. Then I introduced the closed blades of a stronger pair of

scissors, curved on the flat, behind the tumor, and lifted it out of the wound. This produced sufficient free space in the orbit to enable me to remove the little part of the tumor which had been left. I seized it with a pair of strong scissors, had it drawn forward by an assistant, and excised it, always using my left forefinger as a guide. The eyeball, which had been turned aside, was replaced, and the conjunctival wound closed by sutures. Though the tumor was located within the recti muscles, none of them had been cut. The tension of the globe was not changed. The inner half of the eye was anæsthetic, the outer half sensitive, though a little dull. The fundus gave a reddish reflex, but its details could not be seen on account of irregular detachment of the epithelium from the cornea, giving the latter the appearance of frosted glass.

The reaction from the operation was very moderate. The patient now totally lost her headache. On the third day the fundus oculi was visible, and appeared uniformly milky-white. A small whitish-gray infiltration was noticed on the lower part of the cornea. On the fourth day this infiltration had increased, but the fundus could be seen through the clear part of the cornea above it. In the milky surface two dark-red streaks were visible, which had increased the next day, and proved to be the main branches of the retinal veins. Every day new retinal vessels were filled with blood, and in some weeks there was a picture of very marked retinal congestion. (See Fig. 2, Tab. IX., Arch. Ophth. and Otol., 1875.) The fundus had so far cleared up that only the region around the papilla and yellow spot appeared milky, the remainder having its natural color. Gradually this milky appearance disappeared, and then extensive retinal hemorrhage took place, which disappeared slowly, and left the retina and inner part of the choroid atrophic and degenerated. (See Plate X., loc. cit.) The infiltration on the lower part of the cornea, which I have mentioned above, disappeared after the eye had been protected by carefully closing the palpebral fissure. The cornea has never since lost its lustre, and but little change took place in the interior of the eye for two years. Lately, some membranous opacities in the vitreous have appeared. The eye has its natural size, is a little more deeply set in the orbit than its fellow, squints somewhat inward and downward, but has not been the cause of any annoyance. The cornea is bright throughout, though its inner half is still as anæsthetic as immediately after the operation. At this time, two years and three months after the operation, the tension of the eye is diminished, and there is some blood in the vitreous. The patient looks healthy, and there are no symptoms of a relapse.

The tumor was a little above the size of a walnut (30 by 27 mm.), and uniformly hardish. Its base was slightly concave, covered with a thin layer of connective tissue which had united it to the sclerotic. The optic nerve passed through it. The outer sheath of the nerve was well recognizable, though its fibres were more or less separated by the elements of the pseudoplasm. The intervaginal space was a little enlarged, and filled with the tissue of the growth. This, when well hardened in Müller's fluid, was uniformly finely granular. It had no capsule proper, but was loosely covered with the adipose tissue of the orbit. Under the microscope, the growth showed a typical specimen of *scirrhous cancer*. A stroma of fasciculated connective tissue inclosed alveoli filled with epithelial cells, which had no more intercellular substance between them than was necessary to cement them. In the neighborhood of the nerve, a limited number of arenoid bodies (sand-globules) were seen embedded in connective tissue, yet not numerous enough to give even a part of the tumor the character of a psammoma. The whole growth was very vascular, and a good part of its stroma had undergone colloid degeneration. The inner sheath was thickened, and infiltrated with colloid and lymphoid bodies, nowhere with cells of an epithelial character. The interfascicular tissue of the nerve was somewhat thickened. In it, and between the nerve fibrils, there were very numerous lymphoid cells, either single or arranged in rows and clusters; *inflammation of the nerve*. The same change was seen in the intra-ocular end

of the optic nerve—a well characterized specimen of *neuritis optica descendens*. For a detailed description of the specimen, and remarks on many interesting points connected with this case, see Arch. of Ophth. and Otol., vol. iv., 1875, pp. 323 et seq., and Plates IX.–XII.

Case VI.—(Described by L. Krohn in Zeh. Klin. Mon., 1872, S. 103–108.) A married woman, thirty years of age, had a tumor the size of a fist in the right side of her abdomen. It had begun six months previously with local pain, which was followed by general debility, emaciation, headache, vomiting, and gradual loss of sight in both eyes. A tumor was also developed in the left side of the abdomen. The eyes, when examined six months after the beginning of the disease, were totally blind, and, ophthalmoscopically, exhibited marked pictures of choked disk, with some retinal hemorrhages. The patient died eight months after the beginning of the disease.

The autopsy showed *carcinomatous tumors* of both ovaries; all the other organs were normal; the cranial cavity in particular contained no morbid formation. There was a small intumescence in each optic nerve near the sclerotic. The intumescence was caused by a new formation in the intervaginal space, consisting of a tissue which was rich in large round and polygonal cells, and had the arrangement of a cancer. The same kind of cells pervaded the inner sheath and the interfascicular connective tissue, and were very numerous around the central bloodvessels. They extended in the nerve and intervaginal space as far as the lamina cribrosa, but did not enter the eye. On the other end, they could be followed a little beyond the optic foramen. The intra-ocular end of the optic nerve was infiltrated with lymphoid bodies.

Case VII.—(Described by V. Szokalski, in Annales d'Oculistique, t. xlv. p. 43, 1861.) A boy of four years, healthy and well nourished, had a considerable forward protrusion of his left eye, which was perfectly movable. His parents attributed this condition to a severe contusion of the temple, received four or five months previously. The sight was good; by ophthalmoscopic examination, the retinal veins were found enlarged. The exophthalmos increased, the eye became very painful and sensitive to light, the lids could not be closed, the cornea grew opaque, the eye blind, and the other eye became the seat of sympathetic irritation. There was no tumor felt between the globe and the orbital walls; but the orbital tissues were a little more prominent and elastic than usual. None of the symptoms of a vascular tumor were present. Szokalski made the diagnosis of *cystic tumor* directly behind the eye, near the optic nerve, within the recti muscles. He extirpated the contents of the orbit. After having enlarged the palpebral fissure and made a circular incision around the globe, close to the orbital wall, he felt with the finger a hard, nodular, and movable tumor behind the globe, which he isolated from its surroundings by incisions along the orbital walls (*rasant les parois de l'orbite*); then he drew it forward with a strong hook, and, with scissors, severed it near the summit of the orbit. A profuse hemorrhage was arrested by placing a piece of ice in the orbit for two minutes.

The eyeball proved to be quite healthy. A centimetre behind the globe, the optic nerve entered into a nodular, transversely oblong tumor, the size of a filbert, through which the nerve passed in the direction of its smaller diameter, like a string through a bead. Its outer sheath passed directly over the tumor, where it became thicker and more adherent. The substance of the tumor was hard, grayish, fibrous, grating under the knife; it presented under the microscope, reticular areolæ, in the midst of which, caudate, uni-nucleated or multi-nucleated cells were accumulated. The nervous fascicles, after their entrance into the tumor, spread like a fan and were imperceptibly lost in its centre; on its other side they reappeared, and entered convergingly into the other end of the nerve. The growth "undoubtedly took its origin in the neurilemmatic partition-walls of the optic nerve, and unquestionably was of the nature of a scirrhous cancer." Though the nerve was divided as near the optic foramen

as possible, its transverse, cut surface showed, by its gray color, that the pseudoplasm had already penetrated into the cranial end of the nerve.

The wound healed so rapidly that the patient was able to leave the hospital at the expiration of three weeks. For almost five years the boy was in good health, but then a local relapse took place. When he presented himself again, the orbit was filled with a hard, somewhat elastic tumor, which projected in the shape of a small apple beyond the distended eyelids. The child was pale, but otherwise healthy, and complained of some lancinating, periorbital pain. The tumor was nowhere adherent to the orbital wall, and, therefore, easily removed. The patient did well enough during the next two days, but, on the third, convulsions and vomiting set in, and he died of meningitis on the sixth day after the operation.

An autopsy revealed congestion of the pia mater at the base of the anterior lobes, and a greenish, sero-purulent exudation at the anterior part of the base of the skull cavity. Under the left anterior lobe, in front of the Sylvian fossa, was a tumor, as large as a nut, growing from the degenerated optic nerve without implicating the chiasm. The adjacent brain substance was softened. The tumor was dense and consisted of a scirrhous substance.

The optic foramen was dilated and filled with a granulated, reddish substance, which connected the cranial with the orbital tumor. The orbit was enlarged to about twice its natural size, mainly at the expense of the ethmoid and maxillary sinuses. The tumor removed from the orbit contained on its posterior side a large cyst, which had been opened during the operation and had yielded about two ounces of a yellow serum. Sections in different directions through the tumor displayed a scirrho-cancerous, lardaceous substance, grating under the scalpel. Some softened portions of it contained a brain-like substance.

I have made a detailed extract of this case on account of its singular clinical and pathological interest. The original and secondary tumors are pronounced scirrhous cancer by the author, yet his description does not fully convince me. The original tumor may have been a fibroma, and the relapsing tumor a myxo-fibroma. According to the author, both tumors were scirrhous cancers, the secondary one having undergone partial softening in its interior.

These seven cases, together with eight others, form the material from which I shall endeavor to frame a general description of the affection under consideration.

Pathologically, the following species of tumor have been observed :—

(1) *Myxoma*, in five cases.

(2) *Fibroma myxomatodes*, in two cases.

(3) *Sarcoma myxomatodes*, in two cases.

(4) *Glioma myxomatodes*, in one case.

(5) *Fibro-sarcoma*, in one case.

(6) *Psammoma* (perhaps alveolar sarcoma or carcinoma), in one case (E. Neumann).

(7) *Scirrhous cancer*, in two cases.

(8) *Secondary cancer*, from primary cancer of ovaries, in one case.

The majority of these tumors consisted either of pure mucoid tissue (five cases, 33 per cent.), or contained a considerable proportion of mucoid substance in addition to their prevalent tissue. In most of these cases the pseudoplasm originated in the inner sheath and the interfascicular connective tissue of the nerve. To these should probably be added the case of scirrhous cancer of Szokalski, the recurrent tumor in which case contained also a large cyst. In three cases (glioma myxomatodes, fibro-

sareoma, and secondary eaneer) the tumors may have spring from the tissue of the subvaginal space and the inner sheath. Two tumors of a carcinomatous structure (Neumann and Knapp) originated in the outer sheath.

Symptomatology.—The age of the patients varied from one and a quarter to forty years. There is no decided preferenee for either eye. The prevalent symptom is exophthalmos, either directly forward, in the direetion of the orbital axis, or showing besides a secondary deviation in accordanee with a greater development of the tumor on one side. The motions of the globe are preserved, but more or less restricted. On palpation, directly behind the globe a softish tumor is felt, the movements of which correspond more or less to those of the eyeball. A free space can be recognized betwcen the tumor and the orbital wall. Neuritis descendens is a eonstant symptom in the early stages, and atrophy of the nerve in the later stages of the affection. The sight is more or less impaired, the field of vision being restricted when the pseudoplasm originates in the inner sheath and perineurium internum. Sight is less impaired, and the visual field remains complete, when the tumor springs from the outer sheath. Intense and persistent pain has been noticed in the cases of cancer (Neumann, Knapp, Krohn); in the other eases there was pain only when the eyeball was inflamed, with the exeeption of one case of myxoma (Gruening) in which intense pain was felt after the protrusion of the eye had existed for four years without pain.

The *course of the disease* shows in all cases a slow but steady increase of the tumor. The longest duration observed was fifteen years (Heymann and Rothmund). First, the posterior part of the eyeball is flattened, and the eye becomes hyperopic; then the eye protrudes, and neuro-retinitis sets in; then opacity, ulceration, and perforation of the cornea occur; after which the coats of the eye are so much pressed together that the optic disk touches the eornea. The ultimate, spontaneous issue eannot be known from the published cases, since the tumor in all was extirpated, and rccovery took place, except in one case in which a relapse implicating the brain occurred, and in which the patient died from meningitis in eonsequence of a seeond operation.

The observations on record are not sufficient to enable us to form a reliable *prognosis* as to the natural eourse of tumors of the optic nerve. All the cases in which the tumor was removed ended in recovery, which, with the one cxeeption of a fatal recurrence, seems to have been permanent. Sight is, of course, destroyed in all cases.

The *treatment*, until lately, eonsisted in the removal of the tumor, together with the eyeball, which operation offers no diffieulty. Two years ago, I sueeeeded in removing such a tumor, and preserved the eye. About one year sinee, Dr. E. Gruening did the same in another ease. Both of these cases have eontinued to do well. In one case (Graefe), the operation was followed by suppurative inflammation in the wound; in another (Szokalski), the operation caused death by meningitis. In none of the other cases was the operation followed by any unpleasant symptom.

In conclusion, I may say that tumors of the optie nerve form, at present, a sufficiently understood chapter of surgery, and that their differential diagnosis is quite possible. Extirpation relieves the patient from disfigurement and pain, and prolongs life. By improved modes of operative proeedure, the tumor may be removed while the eye, which, though blind, is better than a glass eye, is preserved.